Presacral Schwannoma: A Case Report

Presakral Şıvannom: Olgu Sunumu

Presakral Şıvannom / Presacral Schwannoma

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Özet

Şıvannom kapsüllü periferik sinir kılıfı tümörleridir. Presakral bölgede nadiren yerleşirler ve çoğunlukla kadınlarda görülürler. Tedavisi için komplet rezeksiyon zorunludur. Cerrahi rezeksiyon sonrası nüks nadirdir ve genellikle prognozu iyidir. Bu yazıda transabdominal yaklaşımla total olarak eksize edilen 34 yaşında bir kadın presakral şıvannom olgusu sunuldu. Altı aydır takipte olan hastada rekürrens gözlenmedi.

Anahtar Kelimeler

Şıvannom; Pelvik Kitle; Nörilemmom

Abstract

Schwannomas are encapsulated peripheral nerve sheath tumors. They are rarely seen in the presacral area and are reported mostly in women. Complete resection is mandatory for treatment of schwannoma. After surgical resection recurrence is rare, and the prognosis is usually good. A 34 years old woman with presacral schwannoma who is successfully treated via transabdominal total excision is presented here in this report. No recurrence was seen after six months follow up.

Keywords

Schwannoma; Pelvic Mass; Neurilemmoma

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Introduction

Presacral tumors are seen in 1 of 40000 hospital admissions [1,2]. Neurogenic tumors consist of %10-15 of presacral region tumors. Schwannomas account for only a small portion among presacral neurogenic tumors [1]. Because of the rarity of this tumor, we here present a 34 years old female case with presacral schwannoma.

Case Report

A 34 years old female patient who has admitted to a peripheral hospital for menstrual irregularity has been operated by the gynecologist with the diagnosis of myoma uteri. Upon finding out that the fixated presacral mass was not myoma uteri, biopsies were taken and the operation was terminated. The patient was referred to our hospital with the pathologic result of neurogenic tumor. Detailed anamnesis revealed a history of progressive tenesmus. Digital rectal examination revealed a hard, fixed posterior extra rectal mass. The upper limit of the mass could not be reached. Routine laboratory tests and tumor markers were in normal limits.

Ultrasonography showed a solid 80x50mm presacral inhomogeneous mass lying posterior to uterus. Suboptimal CT scan results were obtained due to lack of rectal contrast. Thus CT scan revealed a 7cm rectal soft tissue mass filling the rectal lumen. A 78x76x65mm relatively well defined round mass was detected in the presacral region by the MRI which has similar intensity with the muscle in T1-weighted images and heterogeneous hyperintens lesion in T2- weighted images. After administration of İV contrast dynamic examination showed a more contrasted lesion resembling a neurogenic tumor [Fig. 1]. Rectosigmoidoscopy revealed a bulge on the posterior rectal wall which has normal rectal mucosa. Biopsies also showed normal rectal mucosa.

The patient was operated on through a midline lower abdominal incision. Posterior parietal peritoneum was opened and the tumor was mobilized by sharp and blunt dissection. Without any damage to surrounding tissues, the mass was totally excised [Fig.2]. Minimum hemorrhage from the presacral veins was easily controlled by cauterization. The patient's recovery was uneventful and there was no postoperative neurological deficit.

Macroscopic pathology revealed an encapsulated 7x5x2cm elastic rather hard tissue which has an inhomogeneous sectional surface. ImmunohistologicallyS100 and Vimetidin were positive and SMA was negative. Thus the tumor was reported as Schwannoma. At this case, as a result of six months earlier follow-up period, there was no evidence of recurrence.

Discussion

Schwannomas which are also called as neurilemmoma or neurinoma are a peripheral nerve sheath tumors [3,4]. Although they are the most common type of peripheral nerve neoplasms, they are rarely seen in the presacral region. Trigeminal and vestibulocochlear nerves near the cerbellopontine angle, cervical and brachial plexus, posterior mediastinum, proximal portion of the large peripheral nerves and spinal roots are common sites where schwannomas can be seen[4-7]. Proximal spinal schwannomas can develop neurological symptoms due to compression on the spinal cord. Sacral and presacral schwannomas comprise only %1-5 of spinal schwannomas[4-6]. Presacral schwannomas are mostly seen in women rather than men[6]. Due to their nonspecific presentation and radiographic appearance difficulties are faced in their diagnosis. Because they are slow growing lesions they can reach a large volume without any symptoms when located in a place with large capacity like the presacral region[8]. Many giant schwannoma cases larger than 10 cm are reported[5]. Most retroperitoneal schwannomas are benign. Takatera et al. reported 133 cases of retroperitoneal schwannoma, 96 of which were benign and 37 were malignant[6]. Although presacral tumors may cause nonspecific symptoms, they benign ones are usually asymptomatic. Lesions eroding the sacrum can present with lumbosacral and radicular pain. Depending on the localization of the tumor, urinary disturbances like urinary incontinence or retention, dysaesthetic sensations or paresthesia and atrophy in lower limb scan be seen[1,3,5,7,9,10]. Also perirectal pain, change in defecation habits, sensation of incomplete evacuation, narrowed stools may be seen[8]. Changes in bowel function, urgency and tenesmus were present in our case.

Most of the pelvic lesions in women originate from genital organs. But gastrointestinal, mesenteric, urinary and primary extra peritoneal neoplasms can also mimic gynecologic tumors [8]. Especially tumors in cystic form can be mistakenly diagnosed as ovarian neoplasms. Also schwannomas can easily be mistakenly evaluated as genital tumors. Ultrasonography can be helpful in differentiating solid lesions from cystic ones. CT and MRI are more precise in observing the apparent relationship of the mass to sacral neural foramen and/or to its presumed origin from a nerve [5]. When compared to CT, MRI has more specificity in delineating the tumor from adjacent anatomic structures[2,5,9,10].

Nerve sheath tumors are classified as three types depending on

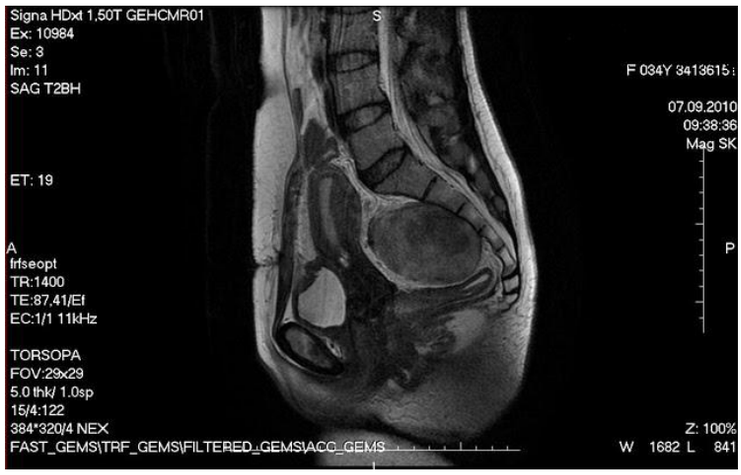

Figure 1. MRI image of the presacral mass.

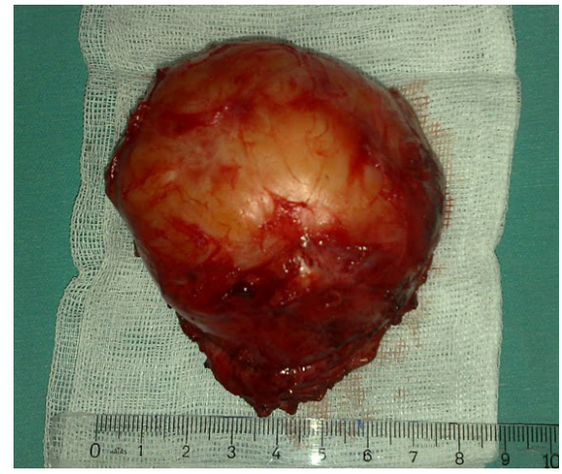

Figure 2. View of totally excised mass.

their localization and spread. Type 1 is localized in the sacrum. Type 2 tumor [most common] is the one which protrudes to adjacent cavities by invading the anterior or posterior sacral wall. Type 3 tumor is localized in the presacral region. In our case tumor was localized in the presacral region [5]. There are also published data revealing cystic schwannomas [3]. When cystic presacral tumors are concerned, differential diagnosis includes anterior sacral meningocele, mullerian ductal cyst, lymphangioma, presacral epidermal cyst and retroperitoneal malignant peripheral nerve sheath tumors [6,7,10]. In our case, imaging studies revealed a solid tumor. Thus, neurofibroma, leiomyoma, malignant tumors of peripheral nerve fibers and malignant melanoma should be considered for differential diagnosis. Nakasima et al. suggested that large tumor size, symptomatic tumor, marginal irregularity and absence of calcifications may predict a malignant tumor [6]. Our case was a large, regular round shaped symptomatic tumor without calcification. It might have been malignant but pathologic result was benign.

Histologically, schwannomas consist of two types which are encapsulated by a true capsule consisting of epineurium. Antoni A type, which is usually predominant, that has highly ordered cellular areas and Antoni B, which has loose myxoid areas. Antoni A type consists of spindle shaped cells with nuclear palisading that are called Verokay bodies. Immunohistologically, diffuse positivity of S-100 protein in the cytoplasm of tumor cells is necessary for definitive diagnosis of schwannoma [1,6,7]. Schwannomas with degenerative changes including cyst formation, calcification and hemorrhage are called 'Ancient Schwannomas'. Ancient schwannomas are rare tumors with long duration usually situated in deep structures such as the mediastinum and retroperitoneum [3,6,8].

Total resection is mandatory for treatment of schwannoma [1,5-8,10]. After complete resection recurrence is rare, and the prognosis is usually good [4]. %10-54 recurrence is reported in incomplete resections like intracapsular enucleation [5,8]. In the presence of recurrence, again surgical resection is necessary [3]. Adjuvant treatment is not recommended even in incomplete resections or recurrences [8]. The choice of surgical approach depends on the size and extension of the tumor. Most authors recommend an aggressive surgery claiming that even loss of bladder and bowel control can be accepted in order to obtain complete excision. Abernathey et al faced a recurrence rate of %54 after 9 years follow-up after enucleation [5]. On the other hand, Dominguez et al. enucleated the tumor on certain cases and have seen %16 recurrence in 9.2 years follow-up [5]. Other surgical approaches to presacral schwannomas are the abdominosacral and posterior approach. A combined abdominosacral approach allows an easier resection of the intrapelvic tumor components and carries low risk of injury to pelvic vasculature [2,10]. Posterior approach offers a better exposure on the nerve roots and caudaequine [5].

Schwannomas are the most common type among peripheral nerve sheath tumors. They can occur anywhere on the peripheral nerve trace but are rarely seen in the presacral region. The symptoms of presacral tumors are similar to rectum tumors and women genital tumors. MRI is the most preferred, precise imaging technique. Both transabdominal or retrorectal approach can be preferred, but the aim should be to resect the tumor totally and complete the operation without any complication.

Competing interests

The authors declare that they have no competing interests.

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